#### **Product Datasheet**

# Recombinant Human Protein ETHE1, mitochondrial(ETHE1)

Catalog No: #AP70283

Package Size: #AP70283-1 20ug #AP70283-2 100ug #AP70283-3 1mg



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# Description

Product Name	Recombinant Human Protein ETHE1, mitochondrial(ETHE1)
Host Species	E.coli
Purification	Greater than 90% as determined by SDS-PAGE.
Immunogen Description	Expression Region:8-254aaSequence Info:Full Length
Other Names	Ethylmalonic encephalopathy protein 1Hepatoma subtracted clone one protein;Sulfur dioxygenase ETHE1
Accession No.	O95571
Calculated MW	43.1 kDa
Tag Info	N-terminal 6xHis-SUMO-tagged
Target Sequence	VARRQLSQRGGSGAPILLRQMFEPVSCTFTYLLGDRESREAVLIDPVLETAPRDAQLIKELGLRLLYAVNTHCH
	ADHITGSGLLRSLLPGCQSVISRLSGAQADLHIEDGDSIRFGRFALETRASPGHTPGCVTFVLNDHSMAFTGDA
	LLIRGCGRTDFQQGCAKTLYHSVHEKIFTLPGDCLIYPAHDYHGFTVSTVEEERTLNPRLTLSCEEFVKIMGNL
	NLPKPQQIDFAVPANMRCGVQTPTA
Formulation	Tris-based buffer50% glycerol
Storage	The shelf life is related to many factors, storage state, buffer ingredients, storage temperature and the stability
	of the protein itself.
	Generally, the shelf life of liquid form is 6 months at -20°C,-80°C. The shelf life of lyophilized form is 12 months
	at -20°C,-80°C.Notes:Repeated freezing and thawing is not recommended. Store working aliquots at 4°C for
	up to one week.

## Background

Sulfur dioxygenase that plays an essential role in hydrogen sulfide catabolism in the mitochondrial matrix. Hydrogen sulfide (H2S) is first oxidized by SQRDL, giving rise to cysteine persulfide residues. ETHE1 consumes molecular oxygen to catalyze the oxidation of the persulfide, once it has been transferred to a thiophilic acceptor, such as glutathione (R-SSH). Plays an important role in metabolic homeostasis in mitochondria by metabolizing hydrogen sulfide and preventing the accumulation of supraphysiological H2S levels that have toxic effects, due to the inhibition of cytochrome c oxidase. First described as a protein that can shuttle between the nucleus and the cytoplasm and suppress p53-induced apoptosis by sequestering the transcription factor RELA,NFKB3 in the cytoplasm and preventing its accumulation in the nucleus.

## References

Identification of new mutations in the ETHE1 gene in a cohort of 14 patients presenting with ethylmalonic encephalopathy. Mineri R., Rimoldi M., Burlina A.B., Koskull S., Perletti C., Heese B., von Dobeln U., Mereghetti P., Di Meo I., Invernizzi F., Zeviani M., Uziel G., Tiranti V.J. Med. Genet. 45:473-478(2008) Research Topic: Cell Biology

Note: This product is for in vitro research use only and is not intended for use in humans or animals.